Report of the Review of Events around suspected Creutzfeldt-Jakob Disease (CJD) in Nova Scotia

Prepared for the Honourable Angus MacIsaac
Minister of Health, Nova Scotia

August 3, 2004

QUEEN ELIZABETH HOSPITAL

QEH

PRINCE EDWARD ISLAND

August 3, 2004

Hon. Angus MacIsaac
Minister of Health
NS Department of Health
Halifax NS

Re: CJD Review

Dear Hon. Minister:

As Chair of the Nova Scotia Joint CJD Review, I am pleased to forward to you the Committee's final report.

The report represents a thorough analysis of the system response to the CJD events of April and May relative to national standards and/or good practice. Through this analysis, process strengths and process weaknesses were identified. Based on the process weaknesses, recommendations for improvement have been developed for your consideration.

As Chair, I would like to express my appreciation to all of the agencies involved in this joint review. Their openness and unreserved willingness to share from this experience was critical to the completion of the review.

I appreciate the confidence you have shown in me by offering me the opportunity to chair this review.

Respectfully submitted,

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Report of the Review of Events around suspected Creutzfeldt-Jakob Disease (CJD) in Nova Scotia

occurring April - May 2004

1.0 Introduction

On April 28, 2004, Health Minister Angus MacIsaac announced a review of factors associated with possible exposure to Creutzfeldt-Jakob Disease (CJD) through medical procedures at the Queen Elizabeth II Health Sciences Centre (QEII) and Yarmouth Regional Hospital (YRH).

During April and May 2004, two patients were being investigated for symptoms which may have been due to a number of conditions including classic CJD, a rare but uniformly fatal neurologic disease that affects the brain and the rest of the central nervous system. These patients underwent medical procedures which are associated with potential risk of transmission to other patients through hospital equipment.

There is, on average, one confirmed case of classic CJD in Nova Scotia every year. Classic CJD (not the same as variant CJD or the human form of mad cow disease) is classified as spontaneous, familial or iatrogenic. Spontaneous CJD, which accounts for 85% to 90%[1] of all cases, occurs apparently spontaneously in the general population with no identifiable cause. The familial form accounts for approximately 10% to 15%[2] of all cases where the family has some history of the disease. The least common iatrogenic CJD, accounting for less than 1%[3] of all known cases, is the result of exposure to medical treatment. Because prions, the protein particles which transmit CJD, resist all routine sterilization and disinfection procedures commonly used by patient facilities, there is a remote risk of transmission of CJD from one patient to another through medical equipment. Not all human tissue is considered to contain prions which have the potential to transmit CJD.

Measures were taken by both Capital Health and Southwest Health[4] to remove the medical equipment used on the two suspect cases from use and to notify other patients who may have been exposed to classic CJD (contacts) prior to isolation of the equipment. In addition, the public was informed of the events which had transpired and the actions being undertaken to prevent further potential transmission of classic CJD.

Through media reports and debate in the Nova Scotia Legislature, public concerns were raised over the adequacy of procedures in place at one hospital and the timeliness of public communication about the events there. This media attention caused a family and community in southwest Nova Scotia to speculate that this was their family member who had been a recent patient at YRH. Southwest Health staff pursued this, which led to medical equipment being removed from service and potential contacts of a second case being notified.

In both cases, risk of contamination with instruments exposed to CJD was later ruled out. Patients contacted in April were subsequently contacted and informed.

2.0 Purpose

This report outlines the process undertaken in Nova Scotia to review factors associated with possible exposure to CJD through medical procedures at two hospitals during April and May 2004. It presents the findings of a critical examination of process strengths and weaknesses and offers recommendations to strengthen the health system's ability to protect the safety of patients and to communicate on public health risks in an accurate and timely manner. The recommendations are contained in Appendix A.

The review was undertaken for the purpose of improving quality. This is done by identifying lessons learned from experience, sharing these lessons for the benefit of the entire system and taking action.

It is also hoped that this report may contribute to media and public understanding of the transmission risks associated with a complex and rare disease and the need to allow sufficient time to gather and communicate accurate information.

3.0 Review Process

The provincial review of the events around the two suspected cases of Creutzfeldt-Jakob (CJD) in Nova Scotia brought together the organizations involved to review organizational response against guidelines or good practice, build on the lessons learned and make recommendations for action. The Minister of Health committed to releasing the review recommendations in order to inform the public of action required to correct or strengthen health system processes.

Pat Lee, Executive Director of the Queen Elizabeth Hospital in Charlottetown, agreed to serve as objective external chair of the review process. The membership of the review team included representatives of Health Canada, the Nova Scotia Department of Health, Capital Health and Southwest Health (see Appendix B).

The review process delved into the following issues:

- the roles and responsibilities of the individual organizations as they pertain to managing infection control issues associated with CJD
- the nature of interaction required among organizations to fulfill the requisite roles
- mechanisms for communicating within the health system and to the public
- authority documents applicable to the topic

- fundamental requirements needed for any organization to manage the prevention, containment and post-exposure management of CJD
- means by which these requirements were met during April-May 2004
- good practices and gaps in existing processes

As part of standard quality improvement practice, the Nova Scotia Department of Health, Southwest Health and Capital Health each reviewed their own activities with regard to the two cases. The Chair of the review team met with each of the three Nova Scotia organizations separately. In addition, joint team meetings were held to finalize the review process, establish the chronology of events, review relevant procedures, discuss health system issues identified in quality reviews conducted by individual organizations, and develop recommendations for action.

4.0 Background

(*In order to safeguard patient confidentiality, certain details surrounding the chronology of events are omitted.*)

The Joint Review Team's first task was to establish the nature and sequence of events pertaining to the two suspected cases. This was an essential first step in the review and laid the foundation for the subsequent critical analysis of actions relative to guidelines or good practice. The scope of this exercise included the organization's action relative to all phases of the CJD event.

A summary of the major events is provided below:

Situation 1

Patient 1 was admitted to the Queen Elizabeth II Health Sciences Centre (QEII) in Halifax for investigation of neurological symptoms. Among numerous other diagnostic tests, a tissue specimen was sent to the QEII pathology laboratory. The tissue had characteristics which could indicate a number of factors, including CJD. However, the tissue's characteristics were inconsistent with the clinical presentation of the patient. The specimen slide was sent on to a specialized neuropathology lab in Toronto for further expert diagnostic review of the unusual tissue characteristics. The possibility of CJD was deemed to be remote. The Toronto lab provides consultation across Canada and also provides contracted expertise to the Health Canada CJD Surveillance System.

The neuropathology lab in Toronto alerted the Health Canada CJD Surveillance System about the case. After reviewing the findings, Health Canada upgraded the likelihood of classic CJD to a probable diagnosis. As a result, a potential risk of contamination of medical instruments was deemed to exist, warranting quarantine of the equipment by the QEII. Some scheduled surgery was cancelled because the equipment was no longer available.

A comprehensive process of assessing instrumentation for risk of infectivity, identifying instruments and tracing contacts was undertaken by the QEII over three consecutive days. During this time, Health Canada and the Nova Scotia office of the Provincial Medical Officer of Health were consulted on infection control issues. It was determined that three separate opinions to confirm diagnosis should be sought. The Minister of Health was informed of the situation.

Physicians began notifying 26 patients of the possibility of exposure to classic CJD and media releases were issued. A public information line was established by the hospital to handle patient inquiries.

Because the diagnosis of classic CJD was still not definitively confirmed, the specimen slide was sent to a specialist in Europe for another opinion.

The following month the diagnosis of classic CJD was ruled out by analysis of the tissue by a third expert in Geneva. The contacts were notified and a public announcement was issued.

Situation 2

Patient 2 had been admitted to Yarmouth Regional Hospital (YRH) for clinical investigation. At the time of the YRH admission, the possibility of CJD was not considered by clinicians. Patient 2 was later admitted to the QEII for investigation during the same time that patient 1 was there. The patient's attending physician at the QEII indicated to the family of patient 2 that the diagnosis might be CJD. The facility in Yarmouth received questions from the family after they heard the media coverage about patient 1 and assumed it was their relative.

The medical device used on patient 2 was quarantined immediately when it was established that patient 2 had undergone an investigative procedure which involved the use of a piece of medical equipment which could require special precautions if it had come in contact with what is considered " low infectivity"[5] tissue. The Provincial Medical Officer of Health was consulted on infection control issues. The Minister of Health was informed of the situation. A media release was issued. A public information line was established by the hospital to handle patient inquiries.

A process of identifying instruments and tracing contacts was undertaken by the YRH over 24 hours. Officials began to notify 32 patients who had undergone a procedure with the same medical device to inform them of the events which may have led to possible exposure to classic CJD.

After review of the tissue sample which had been in contact with the medical device in question, the pathologist at YRH, officials at Health Canada and the Department of Health, determined that no risk of exposure to CJD from patient 2 had occurred to other patients at YRH. Within three days of the initial public release of information about the situation at the YRH, the contacts were notified of Health Canada's assessment and a media release to that effect was issued.

5.0 Findings

Based on the documented events, the Joint Review Team analysed the systems response (the collective response of the organizations) to the CJD event relative to conformance to national guidelines or good practice. Based on this constructive critical analysis, process strengths and process weaknesses were identified. The process weaknesses were used as the basis for most recommendations pertaining to system change and improvement. In some cases, recommendations were formulated to expand or build on the process strengths.

5.1 Infection Control Measures

The need to consider infection control precautions to prevent the transmission of CJD from a patient to other individuals is based on assessment of both patient and tissue risk (see Appendix C). Iatrogenic transmission is deemed possible only when it is established both that there is a probability that a patient has or will develop CJD and that the patient tissues are infective. The circumstances of the two index cases differed in the following ways:

- Patient 1 was provisionally considered a high-risk patient (once the diagnosis was deemed more than remote) and was known to have undergone a procedure in which medical devices came into contact with high infectivity tissue

- Patient 2 was considered a high-risk patient, but it was unknown whether medical devices had come into contact with low infectivity tissue

CJD is not readily diagnosed as there is no diagnostic test available that uses easily accessible biological tissue[6]. Generally, clinicians rely on the clinical presentation of the patient combined with EEG (electroencephalography) and brain biopsy results, if available, to diagnose CJD. Other disease processes which may cause similar neurological symptoms can usually be ruled out through the use of such techniques as imaging and lumbar puncture. A definite diagnosis of CJD may be established only by neuropathological examination (examination of brain or spinal tissue), often conducted post-mortem[7].

At the QEII, the time between consideration of CJD as a remote possibility and isolation of equipment was ten days. The review team deemed this to be reasonable given the circumstances of the case.

Patient 1 represented a particularly complex diagnostic situation, with numerous factors possibly contributing to the symptoms. CJD is included among a long list of disease processes potentially causing such symptoms. A thorough process of investigation of all possible causes was undertaken.

Regular lab processing, analysis and consultation with the attending physician was undertaken within an acceptable time frame of three days. The tissue characteristics seen in the lab were inconsistent with the clinical presentation of the patient. Such tissue characteristics, which could indicate CJD, might also be produced by numerous causes other than CJD, and this was believed to apply in the case of patient 1. This ultimately was confirmed. The diagnosis was not CJD.

Because the clinicians were interested in contributing to scientific knowledge about unusual tissue changes such as these, the specimen slide was sent by the QEII laboratory to the specialized neuropathology lab in Toronto. In such instances, transportation, processing, and analysis of specimens routinely takes approximately seven to ten days. Action to isolate instruments was taken immediately after the determination of probable CJD was received.

This determination of CJD was unexpected. In cases where clinicians suspect CJD, it would be expected that the hospital's Infection Control (IC) department be contacted at the onset of investigation beyond the initial stages of diagnostic differentiation, at the very latest when a sample was taken for analysis. In this case, the clinical assessment did not lead to suspicion of CJD and so IC did not become involved when the specimen was sent to the Toronto lab.

All parties agreed that this was an extraordinarily complex case, and therefore, a total of three separate opinions to confirm diagnosis should be sought. A second national opinion seven days later, did not concur with the first. The third opinion was sought from a CJD expert in Europe. Again, transportation requirements and case complexity prolonged the diagnostic process. The last opinion definitively ruled out CJD 27 days after the first national opinion was rendered in Canada.

Capital Health moved quickly to notify contacts of the final results of the diagnostic consultation.

A second unrelated patient was undergoing diagnostic tests at the QEII during the same time period. Once this was brought to the attention of officials at the YRH, by a query from the family subsequent to the QEII attending physician raising CJD as a possible diagnosis, YRH staff sought and received clarification from QEII the next day that there was a second suspected case. Infection Control staff was involved immediately. The time between notification of a possible case of CJD involving a medical procedure and isolation of the medical device in question was less than 24 hours. YRH moved quickly to seek confirmation of the risk of transmission and to notify contacts of the results negating such risk. This was accomplished within a period of three days.

Process Strengths:

Rapid response on the part of both Capital Health and Southwest Health in isolating the equipment and in communicating publicly to respond to the community's reaction to perceptions of risk.

5.2 Communication

5.2.1 Inter organizational Communication Coordination around the CJD "Event"

The CJD event posed a significant communications challenge to each of the organizations given the perception of CJD as a public health concern, the notification of patients regarding possible exposure to potentially infected equipment, the debate in the legislature and the intense media coverage. An additional level of complexity was encountered based on the mutual interdependence of each of the organizations for exchange of accurate and timely information.

The effective coordination of communication around such a major event as CJD requires that numerous parties collaborate on public announcements. Contingency management benefits from clear protocols which outline roles and responsibilities, enable rapid mobilization of communications and content expertise, and expedite ongoing assessment of events and communications requirements. In the absence of such a coordinated approach, the potential for fragmentation and inconsistent communication exists. Ultimately, the organizations lose the ability to coordinate the release of information and become reactive to media concerns of the moment. Poor coordination has the potential to damage organizational relationships and erode public confidence.

During the course of the review, all parties commended the leadership and support offered by the team in the office of the Provincial Medical Officer of Health in providing and coordinating information among organizations. However, our review of the events indicates that some significant communication breakdowns among the organizations could have been prevented if a joint communication strategy had been developed.

It was apparent to the review team that the lack of DOH policy with respect to expectations around notification threshold and requirements concerning major patient safety events contributed to the communication concerns. At the time the QEII notified the DOH regarding patient 1, past practice was followed with the QEII Vice President contacting the Acting Medical Officer of Health and the QEII communications staff informing the DOH communications staff of the event. The QEII made the assumption that the information would be transmitted further within the department after the initial notification. The DOH expected a more comprehensive notification process, which would require access to a wider range of senior administrative staff 24/7 .

The CJD events demonstrate that any health care delivery organization might have to contact another to deal with unanticipated situations. It is essential that each organization have an established process to mobilize organizational leadership on a 24/7 basis and that this information be shared within the system so that leadership can be contacted quickly. The DOH,

in its coordinating role, must be accessible to all organizations. Existing emergency on-call mechanisms do not adequately support administrative reporting to and within the DOH.

While a number of communication breakdowns were identified, particularly in the first 48 hours of the first event, a number of positive communication practices also occurred. The QEII sharing notification protocols and scripts with YRH, is an example.

Process Strengths:

The sharing between the QEII and YRH of protocols for notifying contacts.

The coordinating role played by the DOH among the organizations involved.

The availability and expertise of the Provincial Medical Officer of Health team.

Process Weaknesses:

The lack of a system policy specifying the circumstances under which the DOH requires it be notified of a major patient safety event and mechanisms to convey information.

The lack of a system-wide 24/7 organizational leadership on call protocol/mechanism.

Recommendations:

- A system-wide policy should be implemented which specifies the circumstances and requirements under which the DOH expects health care agencies to notify the Department regarding major patient safety events.

- The process for accessing administrative staff of each health care agency and the DOH after regular working hours should be outlined and shared across the health system.

5.2.2 Notification of Patients and the Public

All organizations faced numerous ethical and operational challenges associated with communicating publicly about the suspect cases of CJD. The privacy of personal health information must always be safeguarded, yet even seemingly innocuous information about events has the potential to provide enough detail to identify patients. For instance, public inquiries may not be fully satisfied when details of date, diagnosis, gender and place of residence are withheld.

Through the course of the review, the challenges of educating the public about a disease which occurs spontaneously and whose risk of transmissions is described in highly technical terms of probability and theoretical risk became readily apparent.

There is strong commitment across the health system to open disclosure of the effects of patient management on patients and of risks posed to public safety. Although a balance between the public's right to know about safety risks, the privacy of patients, and the need to ensure accuracy of the information provided is required, it is not always possible to achieve. A national guideline does not exist with regard to the notification of potential contacts and public communication. Practices in other jurisdictions, even within Canada, vary on this issue.

It must be noted that the involved organizations seriously considered ethical issues and conflicting demands when communicating with the public. An ethicist was engaged by Capital Health to lead its team through an ethical decision-making framework for contact notification. An important principle of disclosure is that, in respecting the individual's entitlement to full information about his/her own medical conditions or risk to health and safety, the information be imparted in a factual and caring manner. Another principle is that those most directly affected be informed first. This should be done directly by the clinicians or health care organization, not through the media.

In the Nova Scotia cases, potential risk of transmission applied only to patients coming into contact with certain pieces of equipment. This did not represent a general public health threat (such as risk through disease transmitted in the community) warranting immediate public announcements. The first priority was to inform those patients in the two facilities who may have been exposed to CJD through medical devices and to ensure that no further patients were exposed. Because there was no clinical urgency necessitating immediate notification of contacts[8] and the information was likely to cause distress to individuals and their families, both health authorities determined that the best course of action was to wait until the diagnosis (patient 1) or the nature of tissue in contact with the medical device (patient 2) was established before taking action to notify contacts.

The intended timing for notification was changed rapidly because the number of patients and the time frame was the subject of speculation. The responsibility for addressing concerns about public health risks and confidence in the health care system rest both with the Minister of Health and the health care organizations which deliver care. Media releases were issued both by the Department of Health and Capital Health. Once information was made public through the media, it was critical to provide expedient and factual information about the nature and breadth of risk at question.

The risk, with the fast-tracked release of information which identified time frames of potential transmission risk, is that distress can be created for a larger group of individuals who may speculate about whether or not they had been exposed to CJD through procedures at either of the two facilities. Indeed, the perceived need for information was profound in Southwest Nova Scotia, where members of the community and hospital staff began to request information from the District Health Authority. This situation prompted the decision to notify patients who had potentially been exposed to contaminated equipment (contacts) in the absence of definitive information confirming risk.

Because CJD typically has a long incubation or latency period prior to onset of symptoms (perhaps as long as thirty years)[9] notification about possible exposure to CJD causes longstanding concern among potential contacts who, as they age, may show symptoms which are similar to CJD (for example, rapidly progressive dementia) but totally unrelated to CJD. Our review concluded that as difficult as it is for families who have lost a loved one with possible or probable CJD to consider autopsy, it is important to emphasize the need to diagnose CJD through post-mortem examination, as this is the only way that other patients who may have been potentially infected receive definitive information.

With the collaboration of health system stakeholders, the Department of Health is currently developing a policy on disclosure of adverse events. Issues raised in the two cases of suspected CJD should be addressed in this work. Although intended for use by publicly funded health care delivery organizations, this policy may also provide a vehicle to engage the media in planning communication related to health risks within an ethical framework .

Process Strengths:

The presence of notification protocols developed by the QEII to inform possibly infected patients at risk of CJD transmission from contaminated instruments.

Use of an ethical framework for decision-making concerning pros and cons of notifying patients who may have been exposed to a potentially contaminated instrument.

Process Weaknesses

The lack of system-wide notification policies/protocols regarding risk of transmission.

No clear process for the provincial medical officer of health to require an autopsy of possible/probable CJD cases to protect/promote the public good.

Recommendations:

- A process should be developed for collaborative communication planning when informing the public about actual or theoretical exposure to risk within the health care system.
- Mechanisms for safeguarding confidential patient information in public communications should be clearly outlined in communications protocols of all organizations.
- Current provincial policy development on disclosure (underway) should incorporate the use of a consistent ethical framework for decision-making.

- A collaboration between public health, the medical examiner and neuropathology should be encouraged to obtain timely autopsy information in order to provide definitive information regarding risk to contacts who may have been exposed.

5.3 Surveillance

A surveillance system for CJD exists at both the provincial and federal levels. The provincial system is based on mandatory reporting of cases of CJD[10] to permit tracking of cases for purposes of health protection and to facilitate planning and evaluation of health services. Health Canada's CJD Surveillance System is a voluntary, research-based reporting system. Its goal is to determine if there is any risk of developing CJD as a result of receiving blood or blood products or transplants[11].

Reporting of communicable diseases to the Nova Scotia Department of Health is generally well adhered to by practitioners across the province. Because CJD is rare, a special process for reporting is warranted, but has not been delineated in the regulations or procedures supporting the Health Act.

While the Health Canada surveillance system specifies the case definition for reporting to include "definite", "probable" and "possible" diagnoses (see Appendix D), Nova Scotia's system does not specify the level of diagnostic certainty required for reporting. In this event, there was a lack of clarity as to when in the diagnostic formulation the clinician should report. It appears that with case #1, the clinicians were of the opinion that any possibility of CJD was remote and requested that a biopsy be tested primarily for scientific purposes. Consequently, the clinicians neither informed the QEII Infection Control Department nor the DOH, MoH when the specimen was sent to the reference lab and both were subsequently surprised when learning about the possible CJD case other than directly from the clinician.

During the course of review it became apparent that the organizations were sometimes using terminology (e.g. remote risk) different from that identified in the Health Canada CJD Infection Control Guidelines. This led to some confusion about the degree of risk the user was intending to convey.

There is also inconsistent direction on thresholds for reporting within an organization and to external bodies such as the Canadian Blood Services.

Although a link exists for connecting specimens sent to the specialized neuropathology lab to Health Canada's surveillance system, there is uncertainty with regard to the extent to which the Health Canada surveillance system can share information with the provincial surveillance systems.

Process Strengths:

The national monitoring and provincial tracking systems

The Health Canada CJD Infection Control Guidelines

Process Weaknesses:

The lack of clearly defined thresholds for reporting possible cases of CJD

The lack of consistent use of standardized terminology concerning risk of CJD

Recommendations:

- Processes for mandatory reporting of CJD to the provincial Department of Health should be clearly outlined and communicated to the health system.

- Thresholds for reporting of CJD should be clearly stated (i.e. at what level of diagnostic certainty - confirmed, probable, possible) to the following:
  - the Department of Health
  - the CJD surveillance system (Health Canada)
  - Infection Control practitioners within a facility or District
  - Infection Control practitioners or other clinicians in other jurisdictions
  - the Canadian Blood Services

- Opportunities to share or link information between the two surveillance systems should be explored through the federal and provincial departments of health by pursuing any required legislative or policy amendments.

- Health Canada CJD terminology should be used consistently across the province when referring to cases of CJD as confirmed, probable or possible.

5.4 Guidelines

New knowledge about CJD is periodically assimilated into guidelines by various jurisdictions around the world. Although unanimous consensus does not exist, there is significant direction available on preventing and managing CJD transmission. Health Canada's Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, represent expert consensus on the topic in Canada and were used extensively with the cases of suspected CJD in Nova Scotia. These guidelines are highly valued and relied on by clinicians throughout the health system. For this reason, an expectation exists that these guidelines are as current as science permits[12].

In keeping up with advances in knowledge and medical technology, other jurisdictions may assume disparate approaches to infection control management of CJD. This can create confusion for provincial clinicians who seek broad references to support their decision-making. Through the review, it is evident that there is need for a process to regularly update these guidelines to ensure that the guidelines reflect the state of knowledge of the day.

Both health care provider organizations, but especially Southwest Health, were forced to apply the Health Canada guidelines in a very compressed time frame. Through the development of a checklist of required actions, Nova Scotia organizations would be assisted in applying the guidelines to cases of possible iatrogenic transmission. This would be particularly helpful for assigning responsibilities when multi-stakeholder teams are mobilized, as was the case in April and May.

Because guidelines, by definition, are indications or outlines for policy which should not be regarded as rigid standards, every jurisdiction or organization is expected to adapt their own policies and procedures to reflect unique operational requirements. Where the guidelines are silent, for example related to notification of exposed patients and communication among organizations transferring suspected cases, each organization must research and develop its own policies. Capital Health has completed considerable work in developing its operational infection control procedures related to CJD. One area which created significant challenges for Capital Health, was in tracking all instruments associated with the suspected and contact cases.

The case of patient 2, who had a procedure in one facility and was investigated for CJD at another, highlights the need for relevant information sharing in a timely fashion among care providers and organizations in order to identify risk of patients being transferred within the system.

Process Strengths:

Adherence to Health Canada's Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002 .

Capital Health's work in developing its operational infection control procedures related to CJD.

Process Weaknesses:

The currency of the national guidelines.

The lack of system guidelines for the transfer of patient information when a patient with possible, probable or confirmed CJD is transferred from one facility to another.

Recommendations:

- Operational infection control procedures related to CJD developed by Capital Health should be reviewed for feasibility and applicability to other hospitals in Nova Scotia.

- A provincial checklist should be developed for use when multi-stakeholder teams are mobilized, in order to assign responsibilities for requisite notifications and procedures as outlined in guidelines and regulations.

- A process to regularly update national CJD guidelines to ensure that they reflect the state of knowledge of the day should be enhanced.

- Guidelines outlining the type of information to be provided when patients with suspected or confirmed CJD are transferring between Nova Scotia facilities should be developed.

- The feasibility of developing a uniform tracking system for high risk instruments should be explored for use throughout the province.

5.5 Infection Control Capacity

The Infection Control and Infectious Disease practitioners at Health Canada and within the province's two provincial health care centres (QEII and IWK) can provide valuable advice on infection control to health care organizations in Nova Scotia. These experts are in the best position to stay current on rare diseases and should be part of any health care team which is confronted with assessing infection risk associated with CJD.

The staff of Health Canada's Centre for Infectious Disease Prevention and Control as well as the Infectious Disease experts at Capital Health were most helpful in advising Nova Scotia organizations during the events of April and May. Their availability and means of access is not universally known to all organizations. In this case, the Department of Health was able to act in a coordinating role. Consistent with the emphasis on infection control across Canada, the department's capacity to provide a provincial resource and connections to infection control expertise across the system has been undergoing review with a view to further development.

Although the assistance of Health Canada expertise was repeatedly acknowledged during the course of the review, it was noted that this expertise was not accessible 24/7. In case #1 for example, the Infectious Disease specialist at the QEII was able to make off-hours contact with Health Canada expertise in the initial hours of the event through personal contacts rather than through a formalized on-call system.

Process Strengths:

The infectious disease expertise at Health Canada and the QEII.

Process Weaknesses:

Lack of system awareness of the access protocols to infectious disease expertise at the two provincial health care centres (IWK and QEII).

Lack of formalized 24/7 access protocol to Health Canada Infectious Disease expertise.

Recommendations:

- The process for all Nova Scotia hospitals to access infectious disease expertise at the two provincial health care centres (IWK and QEII) should be outlined and shared across the health system.
- The process for accessing infection control expertise from Health Canada after regular working hours should be outlined and shared across the health system.
- The review of Department of Health capacity to provide a provincial resource as well as a coordinating role in infection control across the system should continue to be supported with a view to further development.

5.6 Laboratories

Laboratory expertise in the diagnosis of CJD exists at various centres throughout Canada and around the world. Although transportation of specimens takes time, these expert resources and strong networks enable thorough investigation and diagnostic consultation.

It had not been understood by clinicians at the QEII that when they sent the specimen to the neuropathology lab in Toronto, it would automatically trigger the involvement of Infection Control staff at Health Canada. This happened because the lab which does the surveillance for Health Canada does not differentiate its role in clinical consultation (as in Case #1) from surveillance monitoring. Consequently, any specimen that is sent to the lab is automatically reported to the national surveillance system even if such reporting was not the intention of the sender.

During the course of events surrounding case 1, questions were raised about a possible communication breakdown involving the QEII lab leading to a delay in initiating appropriate infection control measures. The review paid particular attention to the actions taken in the QEII lab. Communication between the lab and the responsible physician occurred as would be expected. As outlined earlier, action to isolate instruments was taken immediately after the determination of probable CJD was received. Our review team is confident that laboratory staff at

the QEII acted professionally and according to established protocols.

Process Strengths:

The international network of expertise.

The expertise of the lab staff at the QEII and other centres.

Process Weaknesses:

Lack of separation at the Toronto reference lab between the roles of clinical consultation and surveillance reporting.

Recommendations:

- The process for clinical consultation with Health Canada's laboratory expertise on CJD should be outlined and differentiated from the voluntary process of reporting cases to the CJD surveillance system, and such information widely disseminated to health care providers.

5.7 Safety Culture

It is commonly understood that, to improve patient safety, the healthcare system must develop, maintain and nurture a culture of safety.[13] This includes openness and frankness in identifying and resolving problems and willingness to share experiences and learn from others. The four organizations involved in the review have participated in a full and positive manner and the lessons learned from this review should be shared provincially and nationally such that the greater system may benefit.

Process Strengths:

The open and constructive critical analysis with which the organizations undertook this joint review such that there could be shared learning.

Process Weaknesses:

Lack of formalized system for sharing information on lessons learned from this event with Provincial and National colleagues.

Recommendations:

- Mechanisms should be sought to share lessons learned from the Nova Scotia experience with all health care agencies as well as other jurisdictions across Canada.

6.0 Conclusion

Every day, Nova Scotia's care providers are faced with clinical cases which are difficult to diagnose and challenging to manage. The expertise and dedication of care providers is commended.

Fortunately, centralized resources are available to provide additional diagnostic opinions and to partner in handling issues of public health safety. In the case of the two suspected CJD cases seen in Nova Scotia during April and May 2004, members of the local, provincial and national infection control team were mobilized and worked together to achieve common objectives.

Findings of the review can be summarized as follows:

- National guidelines were used extensively by all parties.
- Some action is warranted to clarify and strengthen national guidelines and local protocols.
- Action to safeguard against potential exposure to classic CJD was rapid and aligned with commonly accepted thresholds of diagnostic certainty.
- Communication to the public was open and timely.
- Lack of clarity around some communication procedures and the absence of others, served to complicate activity which was conducted under the pressures of public scrutiny and compressed time frames. Communication expectations and mechanisms among the organizations could be clarified and improved.

There are lessons to be learned from the experience. Recommendations for improvement, some extending beyond the management of CJD and more generalizable to organization and communication around all critical patient safety events, are offered for consideration by the Minister of Health.

Appendix A: Recommendations

1. A system wide policy should be implemented which specifies the circumstances and requirements under which the DOH expects health care agencies to notify the Department regarding major patient safety events.

2. The process for accessing administrative staff of each health care agency and the DOH after regular working hours should be outlined and shared across the health system.

3. A process should be developed for collaborative communication planning when informing the public about actual or theoretical exposure to risk within the health care system.

4. Mechanisms for safeguarding confidential patient information in public communications should be clearly outlined in communications protocols of all organizations.

5. Current provincial policy development on disclosure (underway) should incorporate the use of a consistent ethical framework for decision-making.

6. A collaboration between public health, the medical examiner and neuropathology should be encouraged to obtain timely autopsy information in order to provide definitive information regarding risk to contacts who may have been exposed.

7. Processes for mandatory reporting of CJD to the provincial Department of Health should be clearly outlined and communicated to the health system.

8. Thresholds for reporting of CJD should be clearly stated (i.e. at what level of diagnostic certainty - confirmed, probable, possible) to the following:
   - the Department of Health
   - the CJD surveillance system (Health Canada)
   - Infection Control practitioners within a facility or District
   - Infection Control practitioners or other clinicians in other jurisdictions
   - the Canadian Blood Services

9. Health Canada CJD terminology should be used consistently across the province when referring to cases of CJD as confirmed, probable or possible.

10. Operational infection control procedures related to CJD developed by Capital Health should be reviewed for relevance and applicability to other hospitals in Nova Scotia.

11. A provincial checklist should be developed for use when multi-stakeholder teams are mobilized, in order to assign responsibilities for requisite notifications and procedures

as outlined in guidelines and regulations.

12. Guidelines outlining the type of information to be provided when patients with suspected or confirmed CJD are transferring between Nova Scotia facilities should be developed.

13. The feasibility of developing a uniform tracking system for high risk instruments should be explored for use throughout the province.

14. The process for all Nova Scotia hospitals to access infectious disease expertise at the two provincial health care centres (IWK and QEII) should be outlined and shared across the health system.

15. The review of Department of Health capacity to provide a provincial resource as well as a coordinating role in infection control across the system should continue to be supported with a view to further development.

16. Mechanisms should be sought to share lessons learned from the Nova Scotia experience with all health care agencies as well as other jurisdictions across Canada.

Recommendations which may be conveyed to Health Canada:

1. Opportunities to share or link information between the two surveillance systems should be explored through the federal and provincial departments of health by pursuing any required legislative or policy amendments.

2. A process to regularly update national CJD guidelines to ensure that they reflect the state of knowledge of the day should be enhanced.

3. The process for accessing infection control expertise from Health Canada after regular working hours should be outlined and shared across the health system.

4. The process for clinical consultation with Health Canada's laboratory expertise on CJD should be outlined and differentiated from the voluntary process of reporting cases to the CJD surveillance system, and such information widely disseminated to health care providers.

Appendix B: Terms of Reference

NOVA SCOTIA
Department of Health

Review of Events around Creutzfeldt-Jakob Disease (CJD) in Nova Scotia
April 2004

Background

On April 28 Health Minister Angus MacIsaac announced a review of factors associated with possible exposure to CJD through medical procedures at the Queen Elizabeth II Health Sciences Centre. Shortly thereafter, a second case of CJD was identified with a connection to the Yarmouth Regional Hospital. A joint review, involving the provincial Department of Health, Health Canada, Capital Health, and Southwest Health will be undertaken to establish events and to apply the lessons learned from this experience throughout the provincial health system.

Fundamental to our commitment to continuous improvement of Nova Scotia's health care system is the critical examination of process strengths and weaknesses. All of the district health authorities engage in quality review activities which contribute to ongoing improvement. This provincial review will bring together the organizations involved, build on the lessons learned by each, identify areas of cross-over, and make recommendations for the benefit of the entire health system.

Mandate The Review Team is responsible to:

- review the actions and experience of the organizations involved in two recent cases of suspected CJD

- develop recommendations to improve procedures for preventing and managing exposure to CJD in Nova Scotia

Scope

The review will focus on the lessons learned from activity associated with the two suspected cases of CJD identified at the end of April 2004.

Opportunities for improvement identified within the quality reviews of Capital Health, Southwest Health and the Nova Scotia Department of Health will be examined.

Issues which crossed the jurisdiction of each organization will be examined jointly.

Process A preliminary report will be completed within three weeks of finalizing the review team membership.

The review will be coordinated by the Office of the Provincial Medical Officer of Health.

Recommendations will be presented to the Minister of Health for approval prior to public release.

Confidentiality of information pertaining to individual cases will be maintained at all times.

Membership

An objective external consultant will be appointed by the Minister to chair the review team; this will permit all participating organizations to participate fully and on an equal footing.

The Office of the Provincial Medical Officer of Health will coordinate the review.

The Quality Division of the Department of Health will provide support to the Chair of the Review Team and to the review process.

Review team members will represent:

Independent, external chair

Health Canada

Provincial Medical Officer of Health

Nova Scotia Department of Health - Acute and Tertiary Care Branch

Capital Health

Southwest Health

Members

Health Canada	Dr. Antonio Giulivi Blood Safety Surveillance & Health Care Acquired Infections Division Centre for Infectious Disease Prevention and Control
	Shirley Paton Chief, Nosocomial and Occupational Infections Blood Safety Surveillance & Health Care Acquired Infections, Population and Public Health Branch
	Rolande D'Amour Nurse Consultant Blood Safety Surveillance & Health Care Acquired Infections Division Centre for Infectious Disease Prevention and Control
	Jun Wu Manager, Bloodborne Pathogens Section Blood Safety Surveillance & Health Care Acquired Infections Division Centre for Infectious Disease Prevention and Control
Nova Scotia Department of Health	Dr. Jeff Scott Provincial Medical Officer of Health
	Brenda Payne Director Acute and Tertiary Care Branch
Capital Health	Maura Davies Vice President Planning and Performance
	Dr. Alan Cruess Department Chief, Ophthalmology

Southwest Health	Paulette Babin Vice President, Clinical
	Angela LeBlanc Risk Manager and Manager of Infection Control
	Blaise MacNeil Chief Executive Officer
Support staff	Maria Kuttner Quality Management Support Quality, Emergency Health Services, and Health Protection Branch Nova Scotia Department of Health

Appendix C: Risk Assessment for CJD

Health Canada, Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, p. 16 and p. 18.

The risk assessment for CJD is dependent upon two factors: 1) the probability that a patient has or will develop CJD, and 2) the level of infectivity in the patient tissues.

Patient Risk for CJD	
High Risk Patient	**At Risk Patient***
Diagnosed CJD** Suspected CJD: ** undiagnosed, unusual, progressive neurological disease consistent with CJD** (e.g. dementia, myoclonus, ataxia, etc.)	Only under conditions in which there could be exposure to their high infectivity tissues, including CSF (see table next page): recipients of human dura mater grafts †, corneal grafts ‡, and human pituitary hormones members of families with familial CJD, CSS and FFI ¶

* *The incidence of CJD in Canada does not justify classifying people who have undergone neurosurgical procedures as at risk patients.*

** *All forms of classic CJD (sporadic, familial and iatrogenic), GSS and FFI, excluding variant CJD.*

† *Recipients of dura mater grafts are frequently unaware of having received the graft.*

‡ *Corneal grafts originating in a jurisdiction that requires the graft donor to be evaluated for neurological diseases are not considered a risk for CJD.*

¶ *Familial CJD, GSS and FFI can be determined by genetic testing or may be indicated by the occurrence of two or more cases of CJD, GSS or FFI in the family (parent, child, sibling).*

Tissue Risk for CJD	
Level of Infectivity	**Tissues, Secretions and Excretions**
High Infectivity	Brain, spinal cord, dura mater, pituitary, eye* (including optic nerve and retina)
Low infectivity	CSF**, kidney, liver, lung, lymph nodes, spleen, placenta
No Detected Infectivity	Adipose tissue, skin, adrenal gland, heart muscle, intestine, peripheral nerve, prostate, skeletal muscle, testis, thyroid gland, feces, milk, nasal mucus, saliva, serous exudate, sweat, tears, urine, blood, bone marrow, semen

* *The highest levels of infectivity in the eye are associated with the optic nerve and retina and, to a much lower level, the cornea. It is expected that levels of infectivity for other parts of the eye are low or nonexistent. There is no infectivity associated with tears.*

** *Although CSF is classified as low infectivity and is less infectious than high infectivity tissues, it was felt that instruments contaminated by CSF should be handled in the same manner as those contacting high infectivity tissues in high risk and at risk patients.*

Appendix C: Risk Assessment for CJD

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Appendix D: Surveillance Definitions for Classic CJD

(Health Canada, Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, p. 59-60)

Sporadic Case

Confirmed CJD

- spongiform encephalopathy in cerebral and/or cerebellar cortex and/or subcortical grey matter
  AND/OR
- encephalopathy with prion protein (PrP) immunoreactivity (plaque and/or diffuse synaptic and/or patchy/perivacuolar types)
  AND/OR
- scrapie associated fibrils (SAF)

Probable CJD

- rapid progressive dementia
  AND
- typical EEG
  AND
- at least two out of the following four clinical features: myoclonus, visual or cerebellar disturbances (ataxia), pyramidal/extrapyramidal dysfunction, akinetic mutism

OR

- rapidly progressing dementia
  AND
- two out of the four clinical features listed above
  AND
- duration of illness < 2 years
  AND
- 14-3-3 positivity (in cerebrospinal fluid)

Possible CJD

- rapidly progressive dementia
  AND
- two out of the four clinical features listed above
  AND
- duration of illness < 2 years

Endnotes

1. Health Canada, Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, p. 7

2. Health Canada, Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, p. 7

3. Health Canada, Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, p. 7

4. The district health authorities which have responsibility for operating the Queen Elizabeth II Health Sciences Centre and Yarmouth Regional Hospital

5. Only certain types of tissue are deemed capable of transmitting the CJD prion. These tissues are classified by Health Canada as "high", "low" or "no" risk of infectivity.

6. Even though it is possible to conduct biopsy studies , since the biopsy site may not include affected areas, and this is a highly invasive procedure, brain biopsy is generally not encouraged to make the diagnosis.(Health Canada, Creutzfeldt-Jakob Disease Surveillance System in Canada, 1997, p.13)

7. Health Canada, Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, 2002, p. 8

8. There is no screening test available to detect exposure, a lengthy time exists before onset of the symptoms, and no treatment is available.

9. DuVal, Gordon, "Creutzfeldt-Jakob Disease: The Problem of Recipient Notification", Journal of Law, Medicine and Ethics, 25 (1997), p. 34.

10. The Nova Scotia Health Act and Regulations lists the notifiable diseases and delineates the responsibilities of Physicians and Laboratories for the reporting of communicable diseases. These requirements will be replaced and expanded in the new Health Protection Act which will be proclaimed within the next few months.

11. Health Canada, Creutzfeldt-Jakob Disease Surveillance System in Canada, 1997, p.5

12. Health care professionals are encouraged to contact Health Canada for updated information on CJD.

13. National Steering Committee on Healthcare Safety, Building a Safer System: A National Integrated Strategy for Improving Healthcare Safety in Canadian Health Care, 2002.